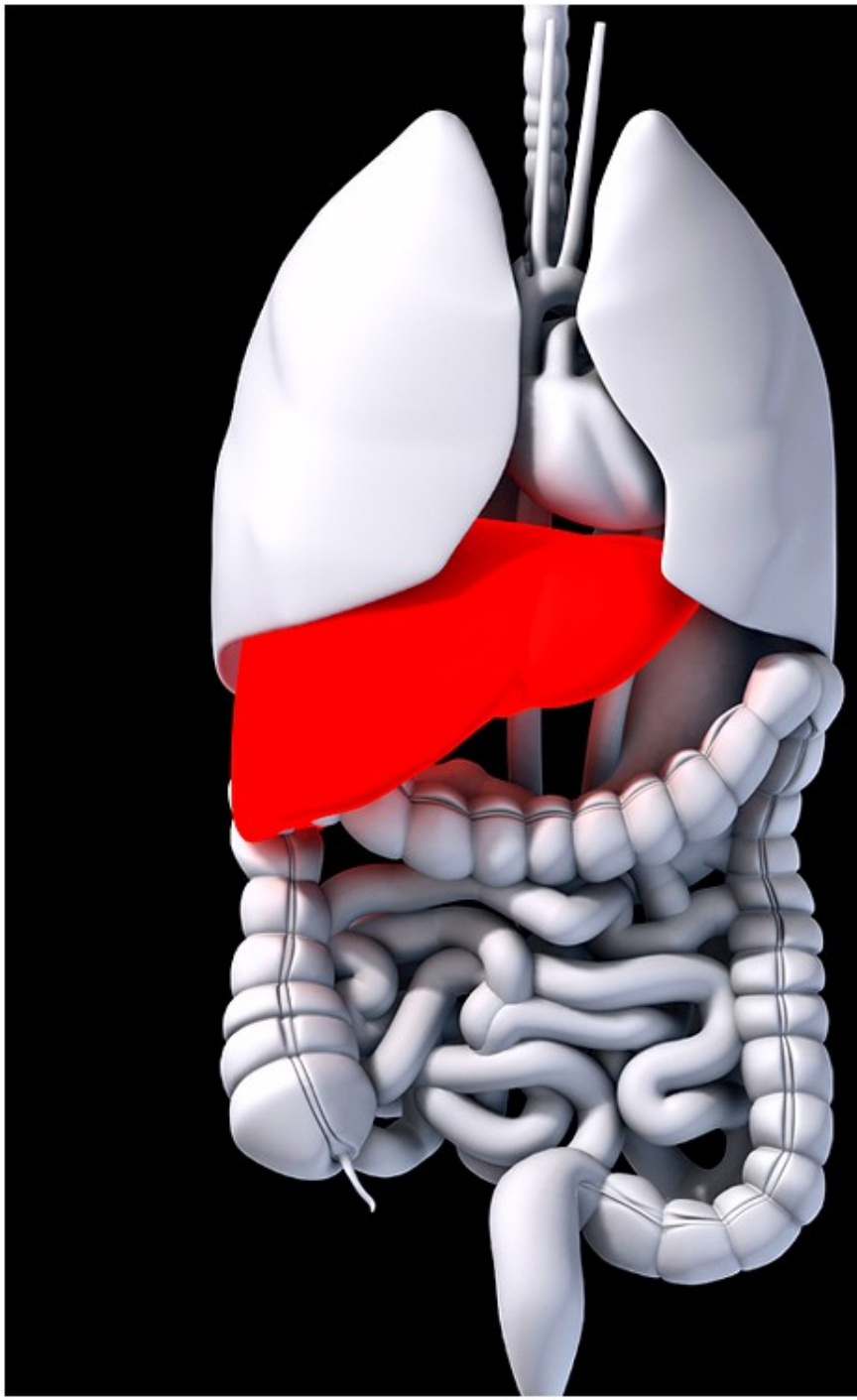
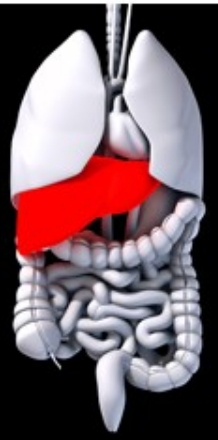


Hirschprung's diseases

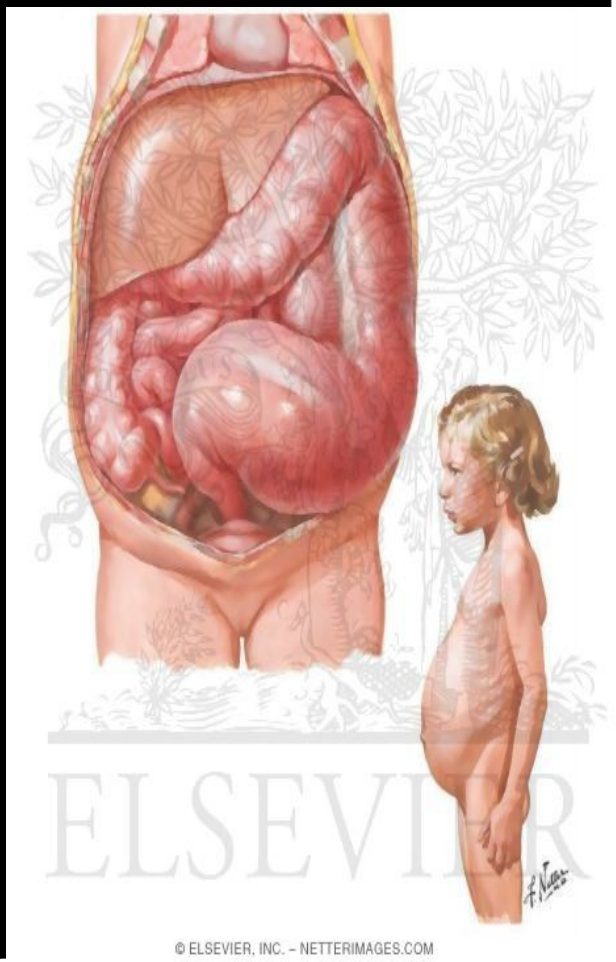
MALAR KODI S
ASSISTANT PROFESSOR
COLLEGE OF NURSING
AIIMS RISHIKESH

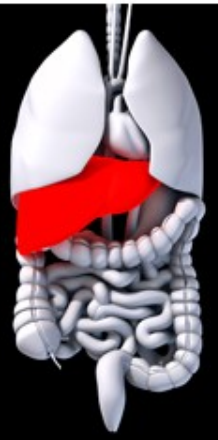


Introduction

Hirschprung's disease called Congenital aganglionic mega colon.

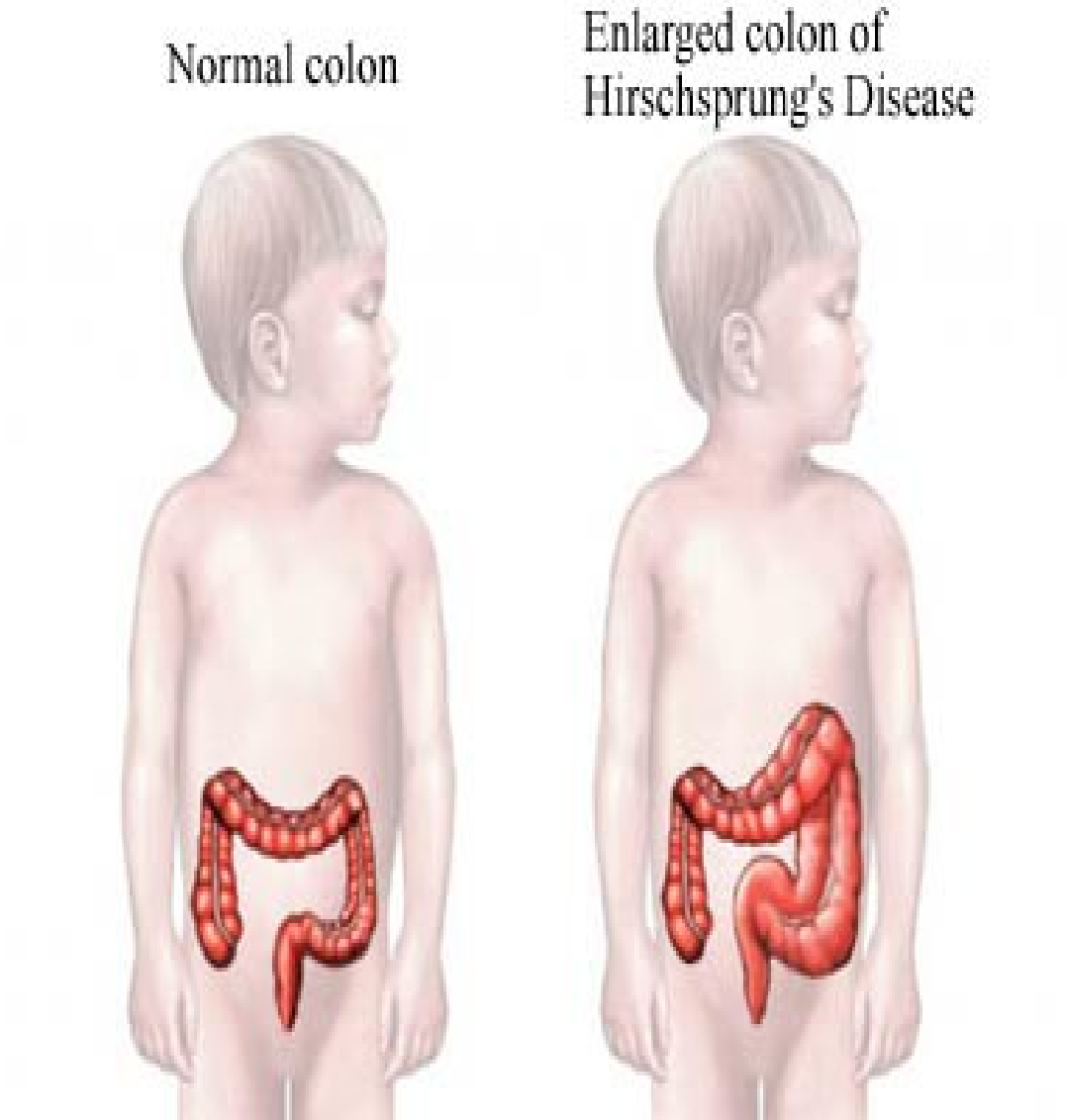
It occurs due to congenital absence of the parasympathetic aganglionic nerve cells, both is muscle layer or sub mucosal layer of distal colon and rectum, which result in extreme dilatation of

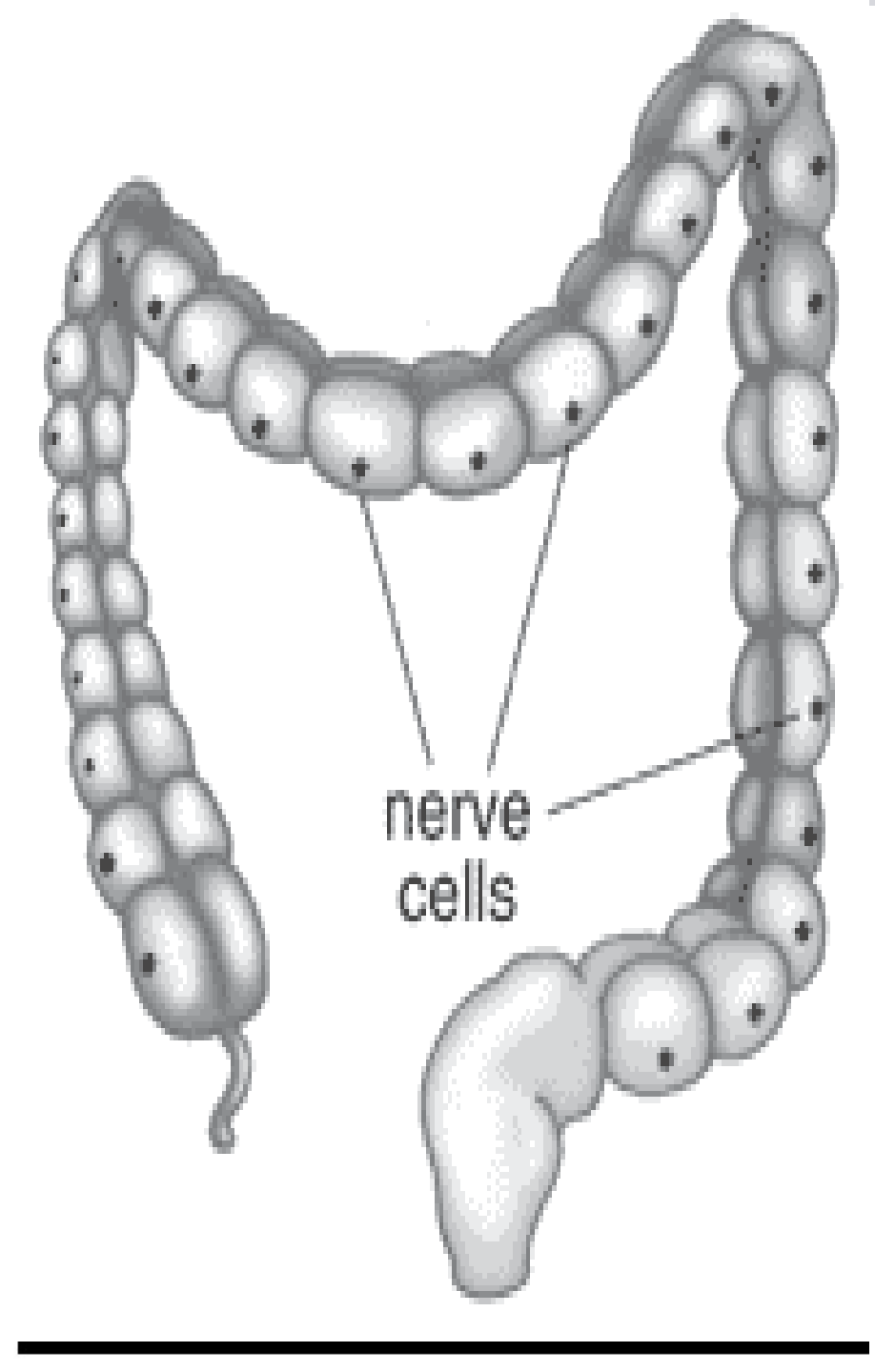


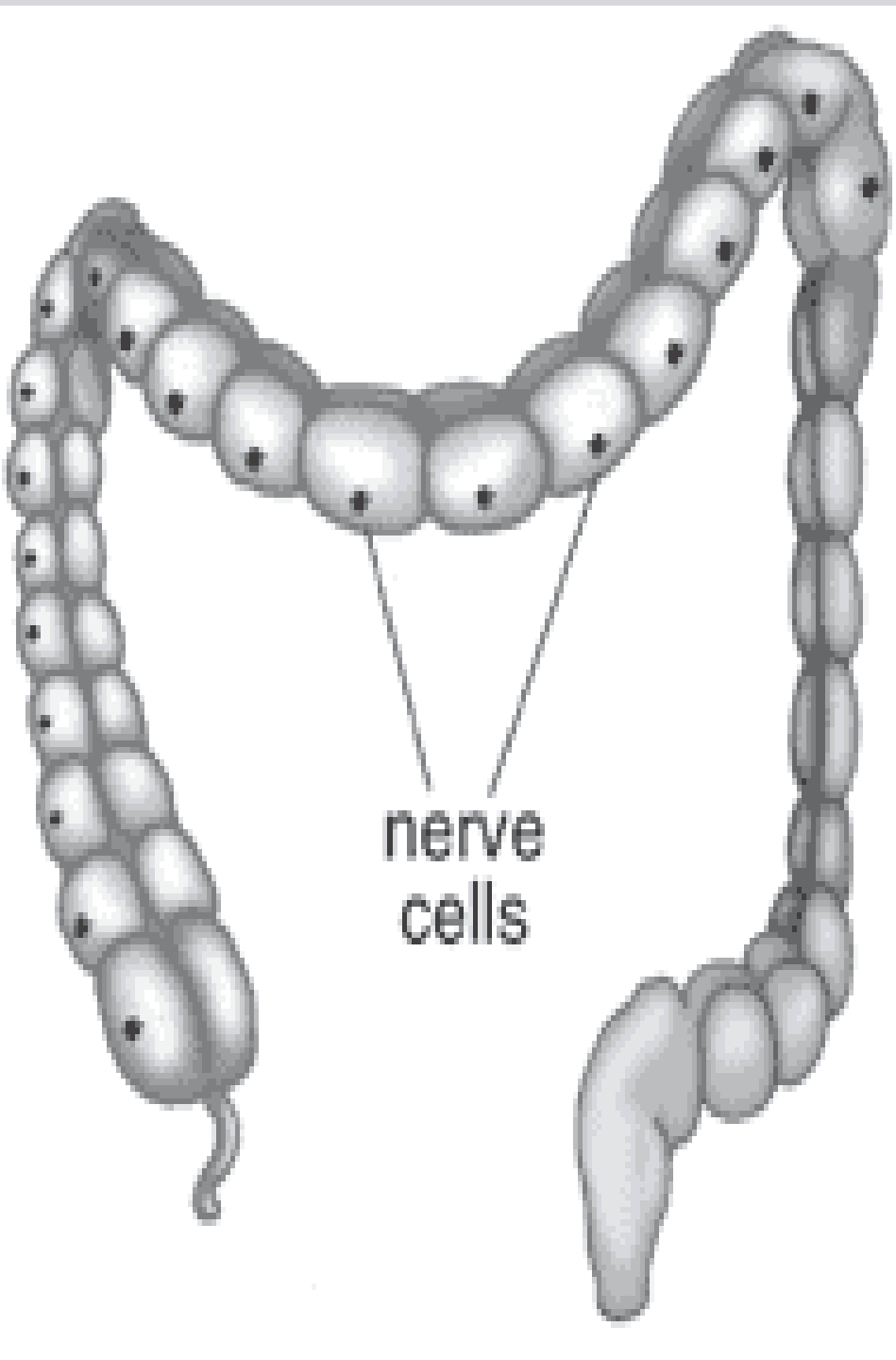


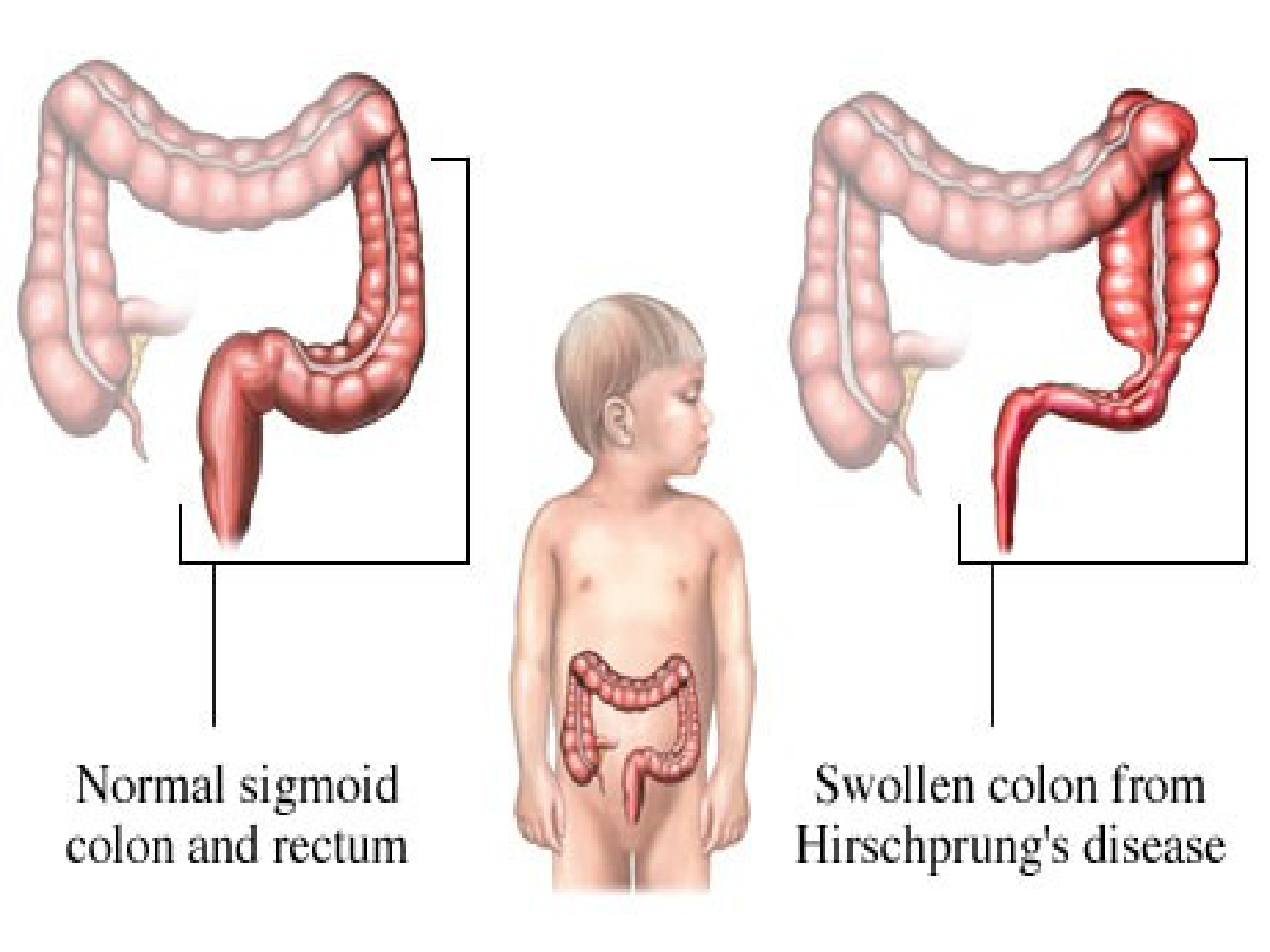
Definition

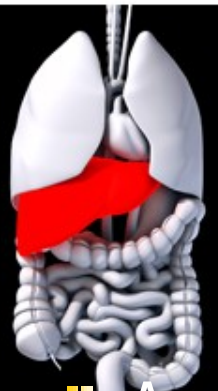
HD is characterized by the Absence of sub Mucosal Ganglion cells in the distal alimentary tract resulting in Decreased Motility in the affected Bowel segment




nerve
cells


nerve
cells

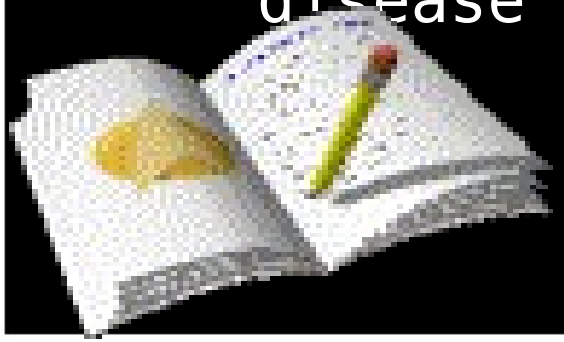

Normal sigmoid colon and rectum
Swollen colon from Hirschprung's disease

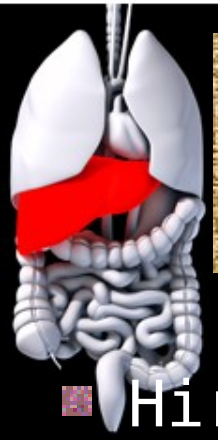


Incidence

- Approximately 1 Per 5000 Live Births.

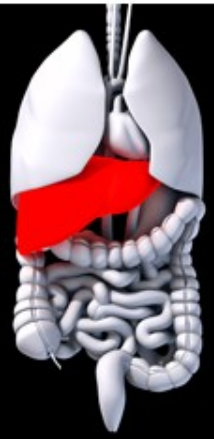
- **Sex:** 4 Times More Common In Males Than Females.
- Nearly All Children With Hirschsprung Disease Are Diagnosed During The First 2 Years Of Life.
- **Mortality/Morbidity:**
  - 25-30%, which accounts for almost all of the mortality from Hirschsprung disease



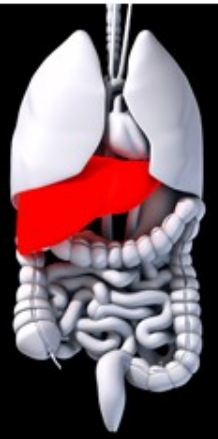


Causes

- Hirschprung's disease develops before a child is born. Normally, nerve cells grow in the baby's intestine soon after the baby begins to grow in the womb. These nerve cells grow down from the top of the intestine all the way to the anus. With Hirschprung's disease, the nerve cells stop growing before they reach the end.
- Causes are unknown
- More come in down syndrome



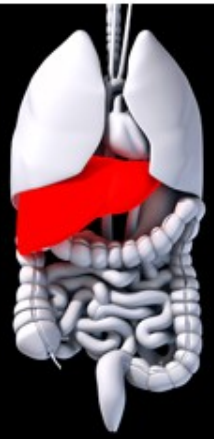
Pathophysiology



Clinical manifestation

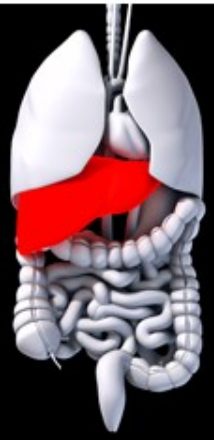
Newborn period:

- Failure to pass Meconium with in 24 to 48 hrs after birth
- reluctance (un willingness) to ingest fluids
- bile stained vomitus
- Abd distention



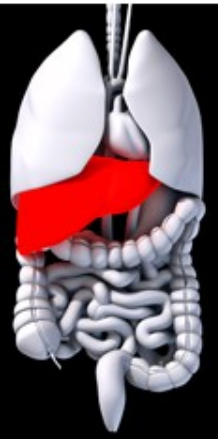
Infancy period:

- Failure to thrive
- Constipation
- Abd. distension
- Episodes of diarrhea & vomiting
- Fever



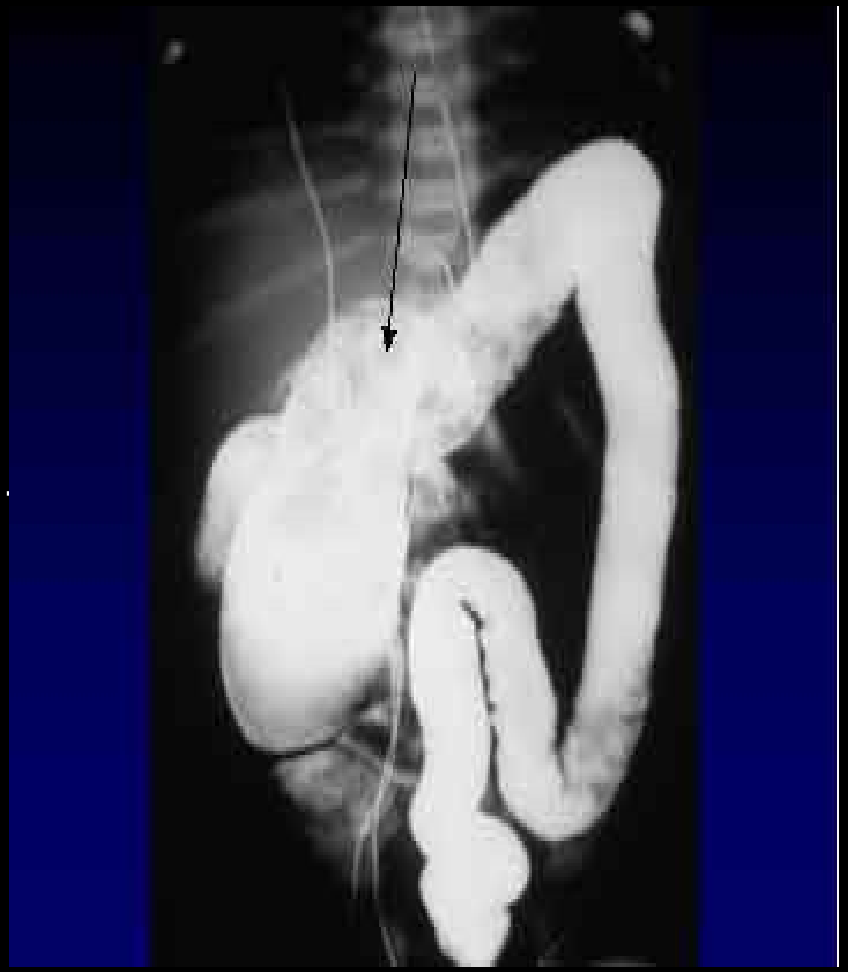
Child hood:

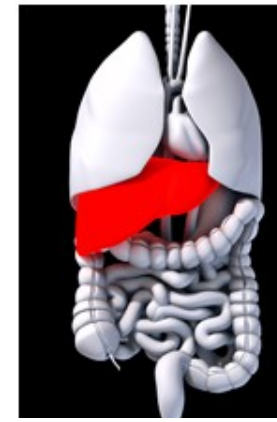
- Constipation
- Ribbon like, foul smelling stools
- Abd. Distension
- fecal masses easily palpable
- Poorly nourished and anemic



Diagnosis

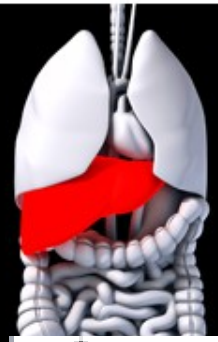
- H.C and P.E
- Radiographic studies
- Barium enema
- Anorectal manometric examination [Absence o normal relaxation of the internal sphincter
- Rectal biopsy – demonstration aganglionic cells in the sub mucosal plexus



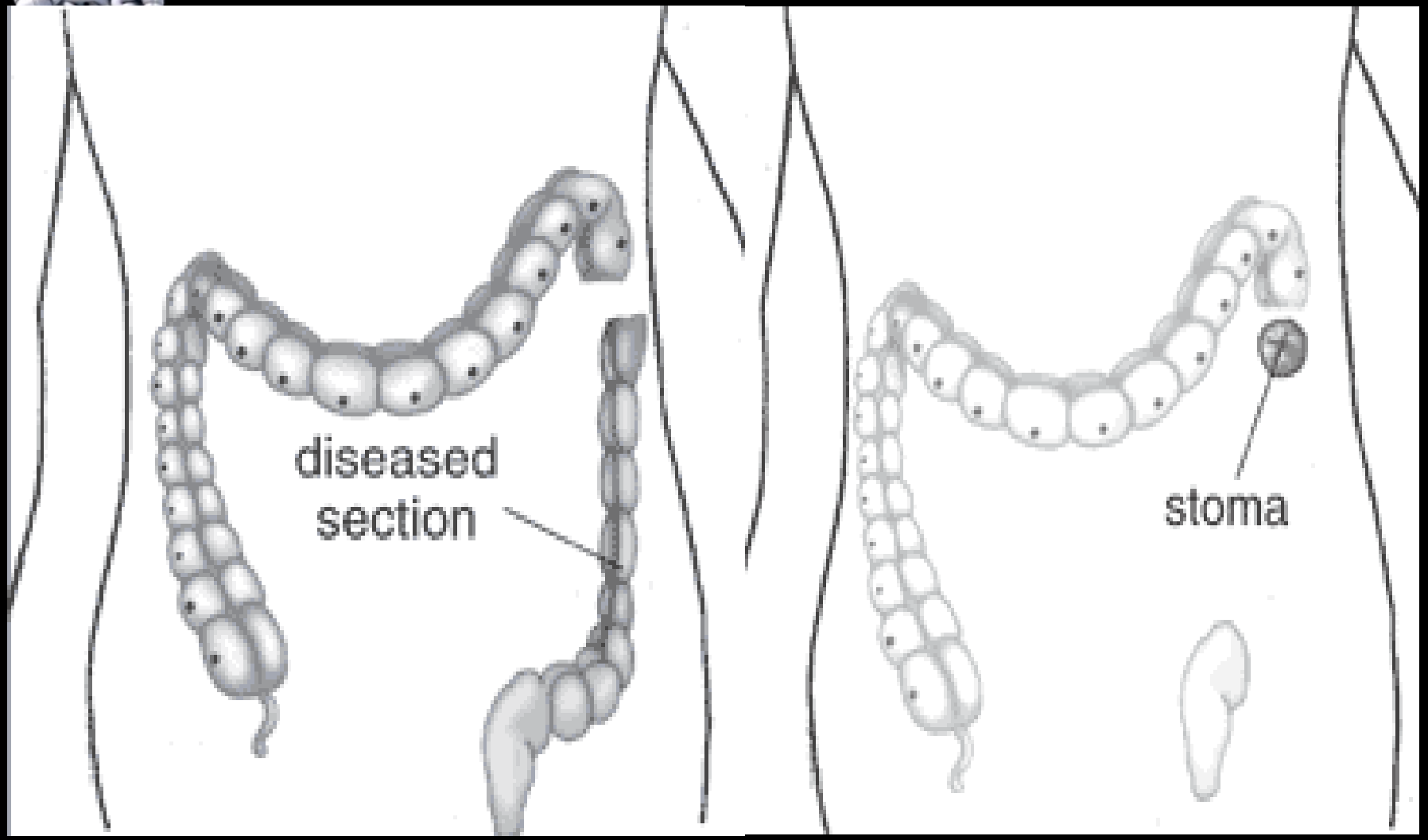


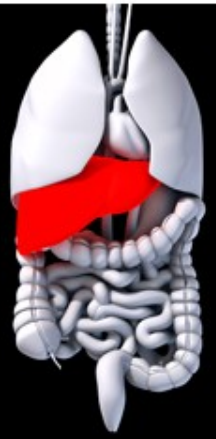
Management

- preliminary colostomy or ileostomy is performed to divert the fecal material and to provide rest to normal colon followed by a definitive pull-through procedure.
- Examples include:
  - Soave pull-through procedure,
  - Swenson procedure
  - Duhamel procedure,



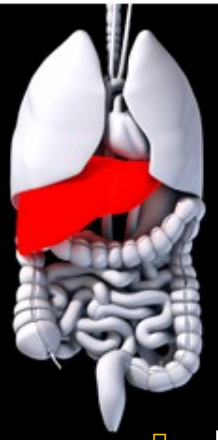
colostomy or ileostomy





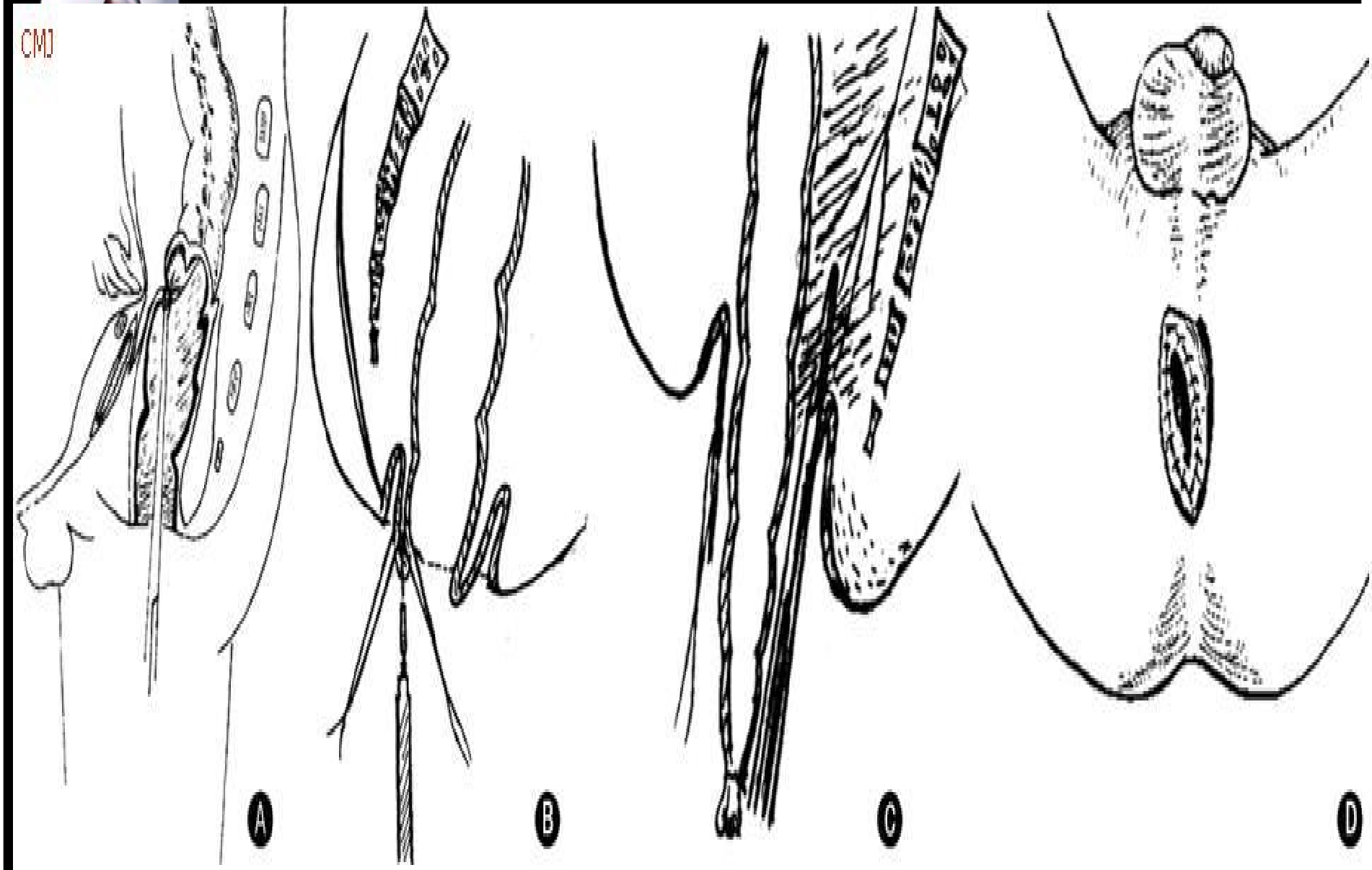
Swenson procedure

- The Swenson procedure was the original pull-through procedure used to treat Hirschsprung disease.
- The aganglionic segment is resected down to the sigmoid colon and the remaining rectum, and an oblique anatomosis is performed between the normal colon and the low rectum.
- Swenson – Up to the sigmoid colon
- Duhamel – Up to the rectum



Duhamel procedure

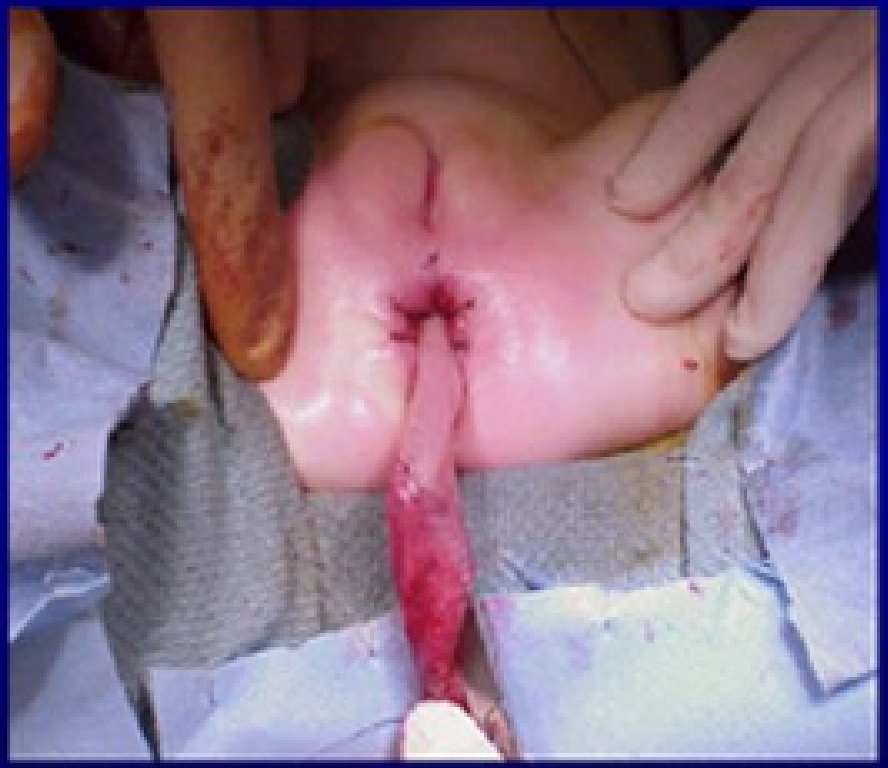
- Duhamel procedure was first described in 1956 as a modification to the Swenson procedure.
- Key points are that a retro rectal approach is used and a significant portion of aganglionic rectum is retained.
- The aganglionic bowel is resected down to the rectum, and the rectum is over seen. The proximal bowel is then brought through the retro rectal space (between the rectum and sacrum), and an end-to-side anatomosis is performed on

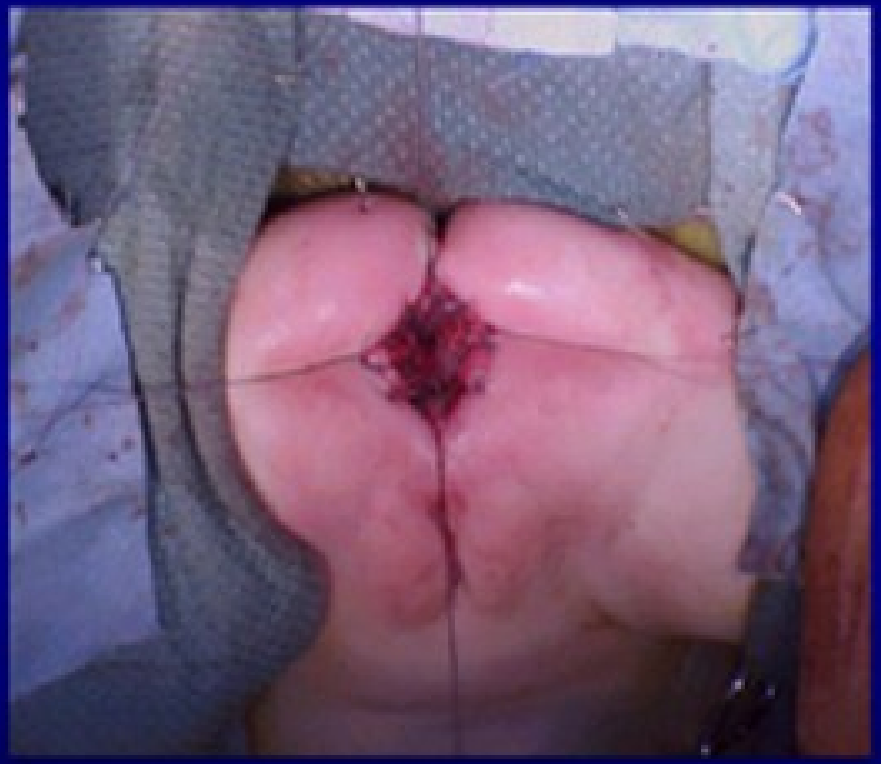

CMJ
A
B
C
D

Hirschsprung Disease

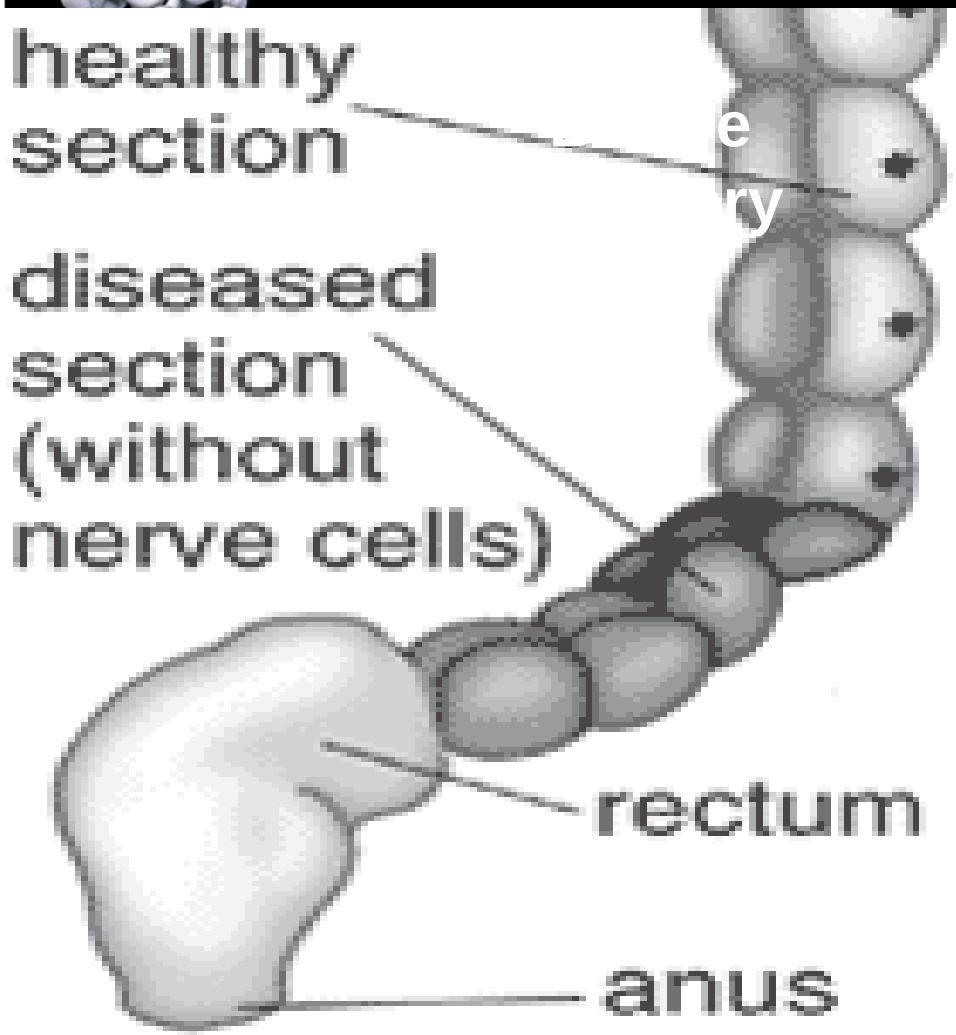
Incisionless Pull through:

Pull through & Anastomosis

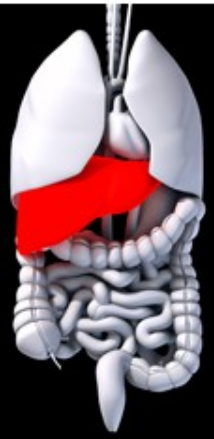




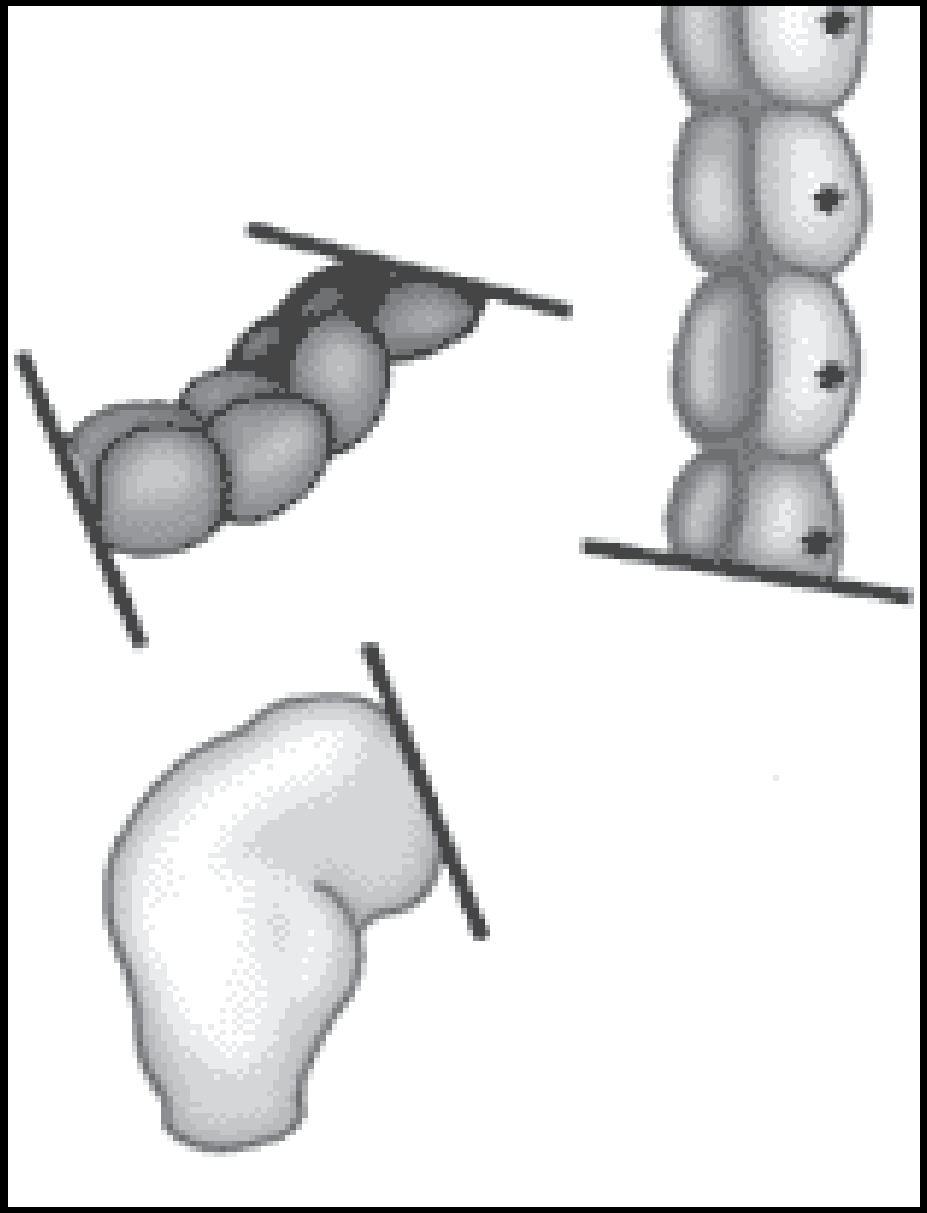
Soave pull-through procedure,

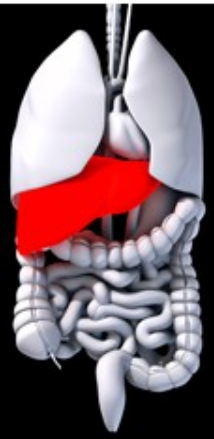


The diseased section is the part of the intestine that doesn't work.

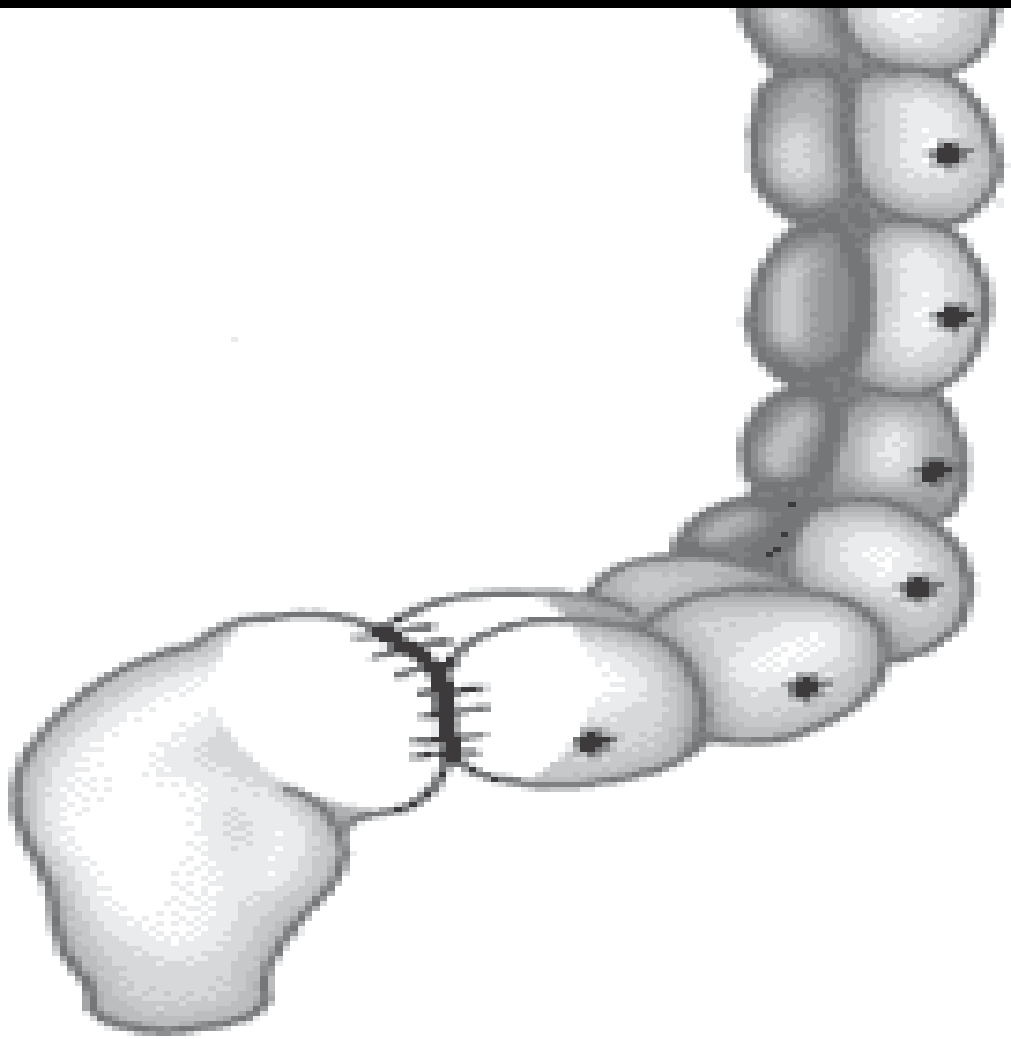


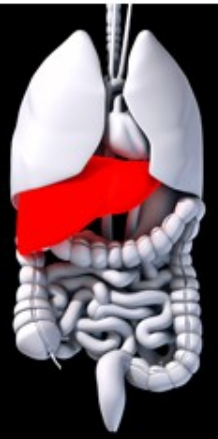
The surgeon removes the diseased section





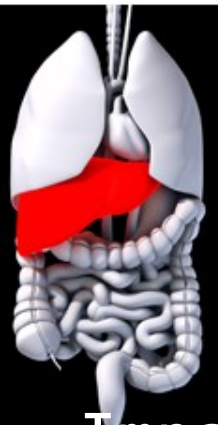
The healthy section is attached to the rectum or anus.





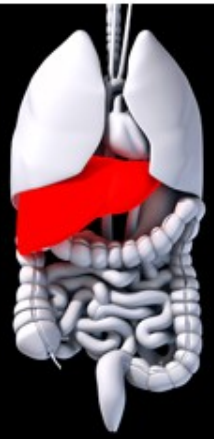
Complication

- postoperative enterocolitis
- anastomotic leakage and stricture formation
- intestinal obstruction
- pelvic abscess
- wound infection and wound dehiscence
- incontinence,
- chronic constipation
- Rectovesical fistulas have also been reported in the literature.



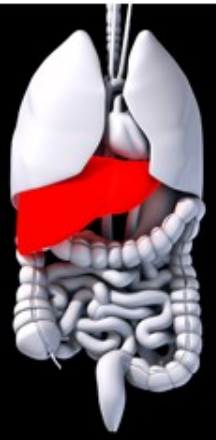
Nursing diagnosis

- Impaired Elimination pattern R/T Aganglionic Segment
- Impaired nutrition, less than body requirement due to poor intake
- Impaired breathing pattern r/t abdominal distention
- Pain r/t intestinal obstruction
- Deficit fluid volume r/t diarrhea and vomiting



- Potential for shock r/t complication like enterocolitis
- Fear and anxiety r/t to life threatening and chronic illness

- Parental knowledge deficit r/t health maintenance in long term illness



Prognosis

- The long-term outcome after definitive repair of Hirschsprung disease
- Unfortunately, approximately 1% of patients with Hirschsprung disease require a permanent colostomy to correct incontinence.
- In general, more than 90% of patients with Hirschsprung disease have satisfactory outcomes, although many patients may have disturbances of bowel function for several years before developing normal continence.

THANKS!